National Reyes Syndrome Foundation UK

TRUSTEE REPORT
EXTRACTS – 30 SEPTEMBER 1989

Significant events

1. Biochemical studies in Children who have had Reye's Syndrome

The receipt by the trustees in September 1989 of a detailed progress report from the Queen's University, Belfast upon completion of a two year investigation titled "Biochemical studies in Children who have had Reye's syndrome.

2. Further studies into the pathogenesis of Reye's Syndrome

In September 1989, the Foundation entered into a commitment with the Queen's University Belfast, not exceeding £29,000 relative to further studies into the pathogenesis of Reye's syndrome.

3. Carnitine Workshop

The Foundation assisted with the financing of a 'Carnitine Workshop' in December 1988 which was held at the Postgraduate Centre, Queen Elizabeth Medical School, Birmingham.

4. BBC Children in Need Appeal

The Charity was able to give help to children who had suffered from Reye's syndrome, largely as a result of a grant from the "BBC Children in Need Appeal".

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APPENDIX
BRITISH PAEDIATRIC SURVEILLANCE UNIT
3rd ANNUAL REPORT
1988-89

Reye's syndrome

Annual totals of reports received for Reye's syndrome (RS) surveillance years 1981/82 – 1987/88 (1 August – 31 July) were: 48, 68, 93, 63, 52, 50, 48. Eighteen were received in the first six months of 1988/89. Of the 48 patients reported in 1987/88, there were 10 who initially met the case criteria but who subsequently had their diagnosis revised (to an inborn error of metabolism in 3). No follow-up information has been received yet for a further six. Of the remaining 32, 17 died giving a case fatality ratio (CFR) of 53%. The median and mean ages were 15 months and 2 years 4 months respectively; there was an excess of males (1.6:1) and there was a winter peak - 14 (44&) cases had their onsets December – February. Apart from the CFR, these features were different from those observed in previous years, when the mean age ranged between 3 and 4 years, the sex distribution was equal and there were no seasonal peaks. Only 10% of patients reported in 1987/88 had a history of pre-admission aspirin exposure, compared to 66% in 1985/86.

Annual total numbers of reports have continued to decline, albeit slowly (although this decline has been striking in N. Ireland which previously had an excess incidence compared to the rest of the British Isles). In spite of better case ascertainment via the BPSU, which was introduced at same time (June 1986) as public and professional warnings were issued about a possible association between RS and aspirin. The dramatic decline between 1985/86 and 1987/88 in cases reported to have taken pre-admission aspirin was not accompanied by a similar sized decline in total number of reports. Earlier concern that RS would be under-diagnosed in the absence of a history of aspirin ingestion brought about by the warnings, was therefore, probably unfounded. There is, however, considerable cause for concern that patients with the inborn errors of metabolism that mimic RS may be being missed. Relatively few of the cases reported have the necessary detailed diagnostic investigations in spite of their very young mean and median ages which make such a diagnosis more likely than "true" RS. The decline in the mean age of RS patients observed since the decline in use of aspirin has also occurred in the USA. It has been suggested that this is due to the increasing proportion of cases that have a biochemical defect caused by the decline in older, aspirin-associated cases.

- **Ideally, all patients presenting with a "Reye-like" illness should be investigated for an in born error of metabolism, but this is especially important in patients under two years of age and/or those with a family history or recurrent episodes. The most important diagnostic specimen is an admission urine taken <u>before</u> any intravenous fluids are given.**

Dr S M Hall - PHLS CDSC

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Cases Reported (Summary)

The numbers of cases reported up to the end of 1988 are shown in table 3. In each column the figure under "A" is the total number of reports received and the figure under "B" is the corrected figure excluding cases not yet followed up, those reported in error and those double-reported within the BPSU system. Numbers of cases given here may differ slightly from the preceding section for reasons of definition and because different time-periods may be used.

Follow-up reports

The time taken to follow-up a report varies greatly between conditions, as does the "accuracy" of reporting measured by the proportion of cases confirmed. Table 4 shows the outcome of follow-up by the appropriate research worker of all cases reported up to the end of 1988. The possible outcomes are explained below the table.

Table 3: Cases reported – 1986, 1987, and 1988 by quarter

	1986 June-Dec		1987 Jan-Dec		1988 Jan–Mar		1988 Apr–Jun		1988 Jul–Sep		1988 Oct–Dec		Total	
CONDITION	A	B	A	B	A	B	A	B	A	B	A	B	A	B
AIDS	25	12	35	10	7	0	19	3	12	2	9	1	47	6
Neonatal herpes	17	8	29	17	8	2	9	3	7	1	3	2	27	8
Reye's syndrome	35	15	45	22	18	8	3	2	9	4	16	8	46	22
Kawasaki	84	72	106	84	25	22	36	24	31	21	37	31	129	98
HUS	35	30	63	47	13	8	20	14	26	23	27	19	86	64
HSES	10	10	17	8	12	5	9	1	7	3	7	2	35	11
SSPE	23	14	27	18	1	1	4	0	8	5	0	0	13	6
Galactosaemia	-	-	-	-	12	4	12	6	7	4	10	3	41	17
Drowning	-	-	-	-	20	18	43	37	34	29	11	9	108	93
IDDM	-	-	-	-	294	266	239	222	302	268	283	226	1118	982
HDN	-	-	-	-	5	2	10	1	12	4	11	6	38	13
ALL	229	161	322	206	415	336	404	313	455	364	414	307	1688	1320

A: All reports received **B: Cases confirmed at 1/4/89**

HUS= Haemolytic Uraemic Syndrome

HSES= Haemorrhagic Shock Encephalopathy Syndrome

SSPE= Subacute Sclerosing Paneccephalitis

Note:- None of the notes set out below the table applied to Reye's syndrome

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Table 4: Outcome of follow-up of cases reported to end of 1988 at 1/4/89

CONDITION	VALID 1a	VALID 1b	INVALID 11a	INVALID 11b	NYK 111	TOTAL	PERCENT IN EACH OF 1	11	111
AIDS	27	21	17	37	25	107	26%	50%	23%
Neonatal herpes	33	0	5	23	12	73	45%	38%	16%
Reye's syndrome	56	3	19	34	14	126	47%	42%	11%
Kawasaki	251	3	24	17	24	319	80%	13%	8%
HUS	98	43	4	8	31	184	77%	7%	17%
HSES	26	3	6	8	19	62	47%	23%	31%
SSPE	22	16	9	2	14	63	60%	17%	22%
Galactosaemia	17	0	4	11	9	41	41%	37%	22%
Drowning	92	1	8	0	7	108	86%	7%	6%
IDDM	970	12	44	26	66	1118	88%	6%	6%
HDN	12	1	0	19	6	38	34%	50%	16%
ALL	1604	83	140	185	227	2239	75%	15%	10%

OUTCOMES:-

1 VALID REPORT:-

1a Case followed up and confirmed by research worker.

1b Case confirmed, but already known to research worker from another source (not a duplicate with the BPSU scheme).

11 INVALID REPORT:-

11a Duplicate report within the BPSU scheme.

11b Reporting error (eg ticked wrong box), revised diagnosis, uncertain case not meeting definition, or unable to follow-up.

111 NOT YET KNOWN:-

Not yet followed up by research worker at 1/4/89.